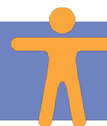
SYSTEMIC LUPUS ERYTHEMATOSUS



SLE is a chronic, multi-system, autoimmune disease with diverse clinical presentation ranging from mild disease characterized by rash and arthritis to a severe disease involving multiple organs.

Pediatric lupus is often more severe than adult lupus. Approximately **20% of SLE cases** are diagnosed in the **first 2 decades of life.**

AMERICAN COLLEGE OF RHEUMATOLOGY (ACR) CLASSIFICATION CRITERIA

Memory Aid *MD SOAP BRAIN*

- **Malar Rash:** fixed erythema, flat or raised, over the malar eminences, tends to spare nasolabial folds
- **Discoid rash:** erythematous raised patches
- **Serositis:** pleuritis or pericarditis
- **Oral/Nasal ulcers:** usually painless
- **Arthritis:** non-erosive arthritis involving two or more peripheral joints
- **Photosensitivity:** rash that develops with UV exposure
- **Blood abnormalities:** cytopenias, hemolytic anemia
- **Renal dysfunction:** persistent proteinuria or cellular casts
- **ANA positive**: doesn't make the diagnosis, helps confirm it
- **Immunologic:** antibodies including anti-ds-DNA, anti-Smith, antiphospholipid antibodies, low complements
- **Neurologic:** seizures or psychosis in the absence of offending drugs or known metabolic derangements

To be classified as having SLE for research purposes, at least 4 out of 11 ACR criteria need to be met. The presence of ≥ 4 criteria is specific for SLE (>93%) and have been used for diagnosis, although not required.

OTHER CLINICAL FEATURES

- Constitutional symptoms, i.e. fever, fatigue, weight loss
- Other rashes, e.g. annular erythema, livedo reticularis, vasculitic rash
- Myalgia and/or myositis
- Raynaud's phenomenon
- Lymphadenopathy
- Hepatomegaly, splenomegaly
- Decreased concentration, cognitive dysfunction, or stroke
- Hypothyroidism
- Pneumonitis, pulmonary hemorrhage
- Pancreatitis, lupus enteritis
- Myocarditis, Libman-Sacks endocarditis
- Alopecia, arthralgias

MANAGEMENT

- **Consult Rheumatology for directed therapy.**
- **Pharmacologic** – Varies based on presentation. Common agents include Hydroxychloroquine (Plaquenil), NSAIDS (If no renal involvement), Steroids (from low dose oral to high dose IV steroids), Disease Modifying Anti-Rheumatic Drugs (DMARDS), biologics, or cytotoxic drugs.
- **Non-pharmacologic** – Sun protection is important to prevent rashes and avoid flares.

INVESTIGATIONS

- **CBCd**: to evaluate for cytopenias
- **Metabolic panel**: may reveal transaminitis, hypoalbuminemia
- **ESR**: can be elevated, CRP: often normal, Ferritin: can be elevated
- **Urinalysis**: screen for proteinuria, hematuria & protein to creatine ratio, elevated creatinine level
- **ANA**: found in 99% of patients with SLE
- **Anti-dsDNA**: very specific for SLE; used to monitor disease activity.
- **Anti-Smith**: highly specific for SLE; found in up to 50% of patients. Other antibodies: SS-A (anti-Ro), SS-B (anti-La), anti-phospholipid antibodies
- **Complement levels C3, C4**: Low in patients with active disease
- ***Annual ophthalmologic exam***
- MRI brain, CXR, ECG, CSF analysis, ECHO as needed

Elevated CRP in patients with SLE can be a sign of infection, serositis, or Macrophage Activation Syndrome (MAS).

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